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Pictorial Essay

Fibrous Dysplasia of the Temporal Bone: Imaging Findings

Eugene W. Brown[1], Cliff A. Megerian[2], Michael J. McKenna[2], Alfred Weber[1]

Lichtenstein [1] in 1938 coined the term fibrous dysplasia to describe a disorder characterized by the progressive replacement of normal bone elements by fibrous tissue. Histopathologically, these lesions consist of an abnormal proliferation of fibrous elements intermixed with haphazardly arranged trabeculae of woven bone. The disease can involve any bone in the body. In the head and neck, the skull and facial bones are involved in 10–25% of cases of monostotic fibrous dysplasia and in 50% of the polyostotic variety. Involvement of the temporal bone, however, is relatively rare, and only 53 cases have been reported. The three major radiographic classifications of fibrous dysplasia are pagetoid, sclerotic, and cystic. Any of these types may involve the temporal bone and related structures, including the external canal, middle ear, jugular foramen, or, rarely, the otic capsule. In this essay, we illustrate the radiographic features of the disease based on our experience with seven cases, seen at our institution since 1977, of fibrous dysplasia involving the temporal bone

Radiographic Features

Fries [2] reviewed the radiographic features of fibrous dysplasia of the skull and facial bones and described three patterns. The pagetoid, or ground-glass, pattern is most common (56%) and consists of a mixture of dense and radiolucent areas of fibrosis (Fig. 1). Sclerotic lesions (23%) are homogeneously dense (Fig. 2), whereas the cystic variety (21%) is characterized by a spherical or ovoid lucency surrounded by a dense bony shell (Fig. 3). Histologic correlates have shown pagetoid lesions to result from an equal mixture of fibrous tissue and woven trabecular bone [3]. This mixture produces radiodense and lucent areas resulting in the classic ground-glass appearance. A predominance of osseous elements results in the opaque sclerotic type of fibrous dysplasia. Similarly, the cystic type results from an abundance of fibrous elements.

CT scanning, the primary mode for radiographically evaluating fibrous dysplasia, is, in our experience, the best way to display the bony changes. Plain films and MR imaging are useful adjuncts. Particularly in cases of cystic fibrous dysplasia, MR imaging may be useful to assess the soft tissue and fibrous components and to evaluate the effect of these primarily bony lesions on adjacent soft tissue structures of the skull base, such as the jugular vein and brain stem.

External Canal Involvement

Involvement of the external canal is the most common manifestation of fibrous dysplasia of the temporal bone, occurring in approximately 80% of patients [4]. Progressive stenosis of the external auditory canal may result, and a conductive hearing loss may occur (Figs. 4 and 5). Clinically, this condition may be misdiagnosed as an osteoma. In addition, canal cholesteatomas, which are thought to be secondary to obstruction, occur in 40% of cases (Fig. 4). Extension to the temporomandibular joint may also occur in some cases, and trismus may be a symptom [4]. Middle ear involvement is uncommon and usually results from long-standing external canal stenosis with secondary cholesteatoma and destruction of the tympanic membrane and ossicles.

Inner Ear and Otic Capsule

Although previous authors [5, 6] have emphasized that fibrous dysplasia does not involve the inner ear structures, it is clear from our experience that the otic capsule may be involved. We

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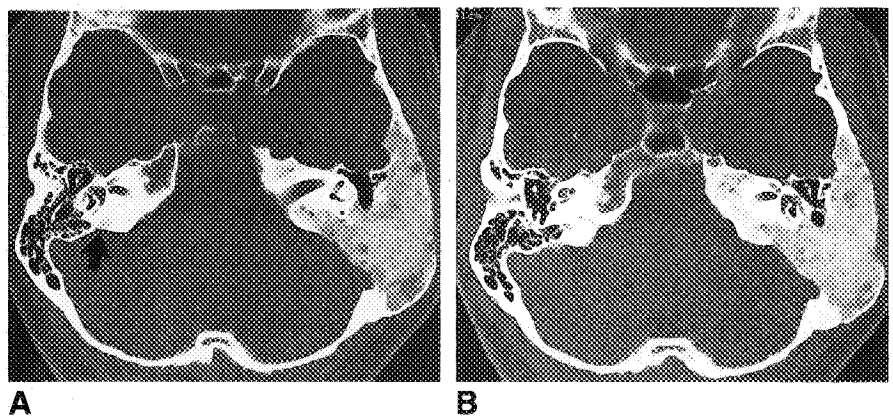


Fig. 1.—33-year-old woman who had CT scan for evaluation of headache. Patient's hearing was normal.

A and *B*, Axial CT scan at level of epitympanic recess shows extensive involvement of left temporal bone and mastoids by pagetoid fibrous dysplasia producing classic ground-glass appearance. Note relative preservation of inner ear structures. Vestibular aqueduct on right is normal (*arrow, A*). Left vestibular aqueduct is obliterated by disease.

C and *D*, Coronal CT scan shows relative sparing of middle ear and otic capsule. Note that disease nearly surrounds superior semicircular canal (*arrow, D*).

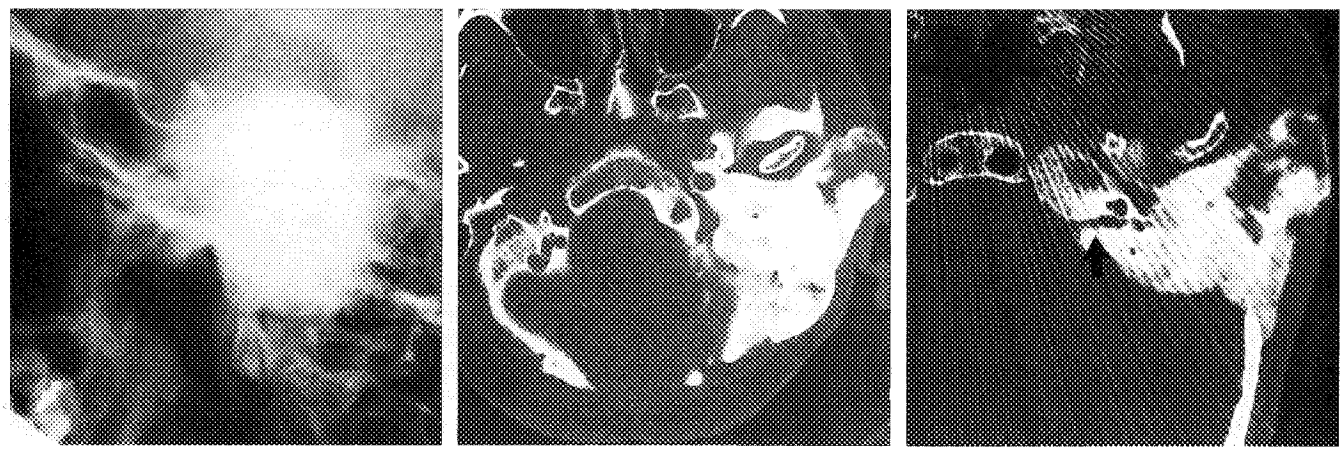


Fig. 2.—37-year-old woman with 10-year history of monostotic temporal bone fibrous dysplasia who developed sudden hearing loss and vertigo.

A, Lateral plain film shows bony overgrowth and sclerosis in temporal bone.

B, CT scan at level of lower mastoid shows extensive bony overgrowth in left temporal bone with predominantly sclerotic appearance. Some lytic areas are in mastoid lateral to temporomandibular joint. Note narrowing of jugular fossa and involvement of carotid canal.

C, Slightly higher section at level of internal auditory canal shows more extensive lytic areas and areas of ground-glass change. Note bony overgrowth causing narrowing of internal auditory canal and bony labyrinth (*arrow*).

have encountered several examples in which fibrous dysplasia involves the cochlea and labyrinth. Sensorineural deafness can result from either cochlear destruction, internal auditory canal stenosis, or vestibular fistulization [4]. The cochlea may ultimately be replaced by the disease process, even in cases that initially are limited to the mastoid and middle ear (Fig. 6). In the cystic type of fibrous dysplasia, the progressive lytic process may erode the vestibule, producing fistulas, vertigo, and hearing loss (Fig. 3). Stenosis of the internal auditory canal and the resulting encroachment of the cochlear nerve may also produce deafness (Fig. 2). According to some authors [5], involvement of the vestibular aqueduct does not occur. However, we observed such involvement in at least one patient (Fig. 1). Involvement of the inner ear may also encroach on the intratemporal facial nerve, and facial paralysis may rarely be seen (Fig. 6).

Temporomandibular Joint

Involvement of the mandibular fossa and middle cranial fossa may also occur in fibrous dysplasia [4]. The bony overgrowth in

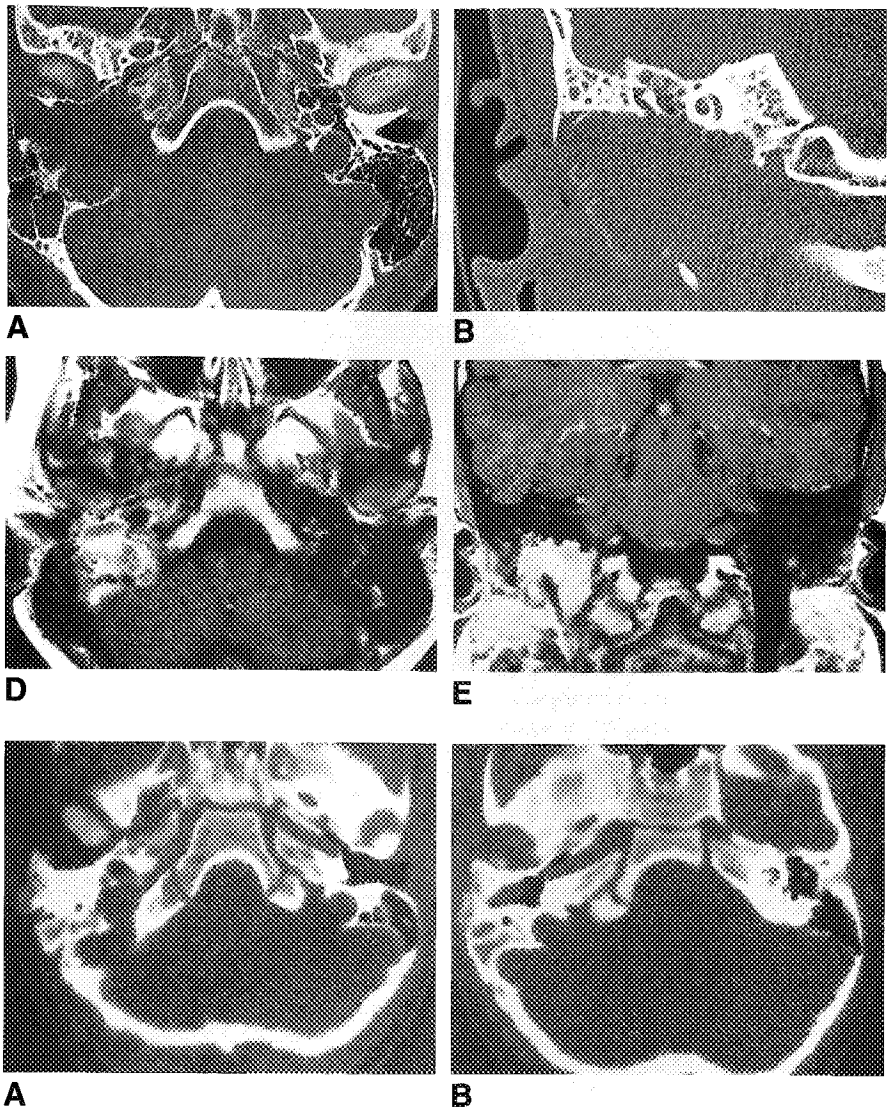


Fig. 3.—9-year-old girl who failed audiogram at school.

***A*, Axial CT scan at level of jugular bulb shows extensive lytic process extending from posterior aspect of temporomandibular joint to jugular foramen, representing cystic type of fibrous dysplasia.**

***B*, Coronal CT image showing erosion of inferior mastoid, middle ear cavity, and otic capsule with erosion of ossicular chain.**

***C*, More posterior coronal image shows evidence of fistula to vestibule (*arrow*). Fistula resulted in severe sensorineural hearing loss.**

***D* and *E*, Axial and coronal postcontrast, short TR images showing enhancing mass in inferior mastoid and in jugular foramen.**

Fig. 4.—4-year-old boy who presented with conductive hearing loss.

***A*, CT scan at level of external canal shows obliteration of right external auditory canal. Note sclerotic appearance with areas of bony overgrowth replacing mastoids.**

***B*, Slightly higher section shows complete stenosis of external canal and soft tissue mass lateral to tympanic membrane. This lesion was cholesteatoma at surgery.**

these areas may deform the temporomandibular joint with resulting decreased excursion of the jaw and trismus (Figs. 2 and 7). Involvement of the middle cranial fossa and displacement of the anterior temporal lobe may occur secondary to disease in the anterior portion of the temporal bone (Figs. 5, 7).

Jugular Foramen

Expansile lesions of the bony jugular foramen may encroach on the jugular vein and lower cranial nerves. Jugular venous thrombosis with retrograde extension to the sigmoid sinus may occur. Cranial neuropathies, especially of nerves IX and X, can result from encroachment of the jugular foramen and fossa (Fig. 7). Bony overgrowth of fibrous dysplasia in this region may extend to compress the posterior fossa and brain stem. Glomus jugulare tumors, chondrosarcomas, hemangiopericytomas, and metastatic disease may involve the jugular fossa and simulate the cystic type of fibrous dysplasia. The cystic type is rare; therefore, any cystic lesion involving the jugular should be fully evaluated diagnostically to exclude these other entities. MR imaging and MR angiography may help in the evaluation by demonstrating vascular lesions that may have a similar lytic appearance on CT.

Differential Diagnosis

The differential diagnosis of fibro-osseous lesions of the temporal bone includes fibrous dysplasia, aneurysmal bone cyst, unicameral cyst, nonossifying fibroma, Paget's disease, osteochondroma, giant cell granuloma, exostosis, osteoma, and ossifying fibroma, as well as sarcomatous neoplasms. The diagnosis of fibrous dysplasia is confirmed with a combination of radiographic and histopathologic data. The presence of woven lamellar bone and a fibrous matrix suggests differential features.

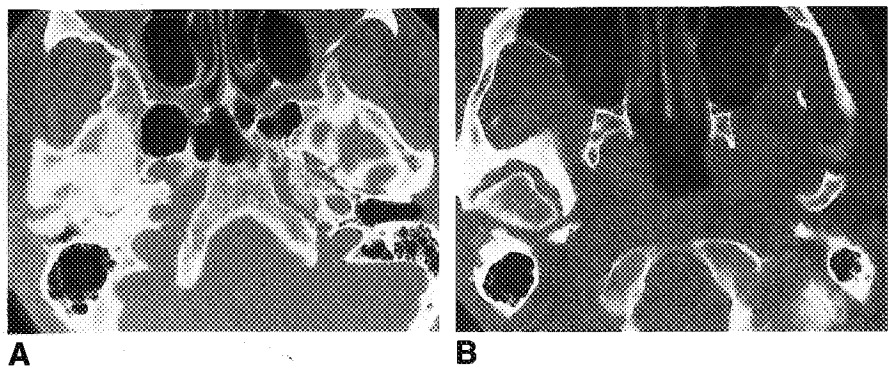


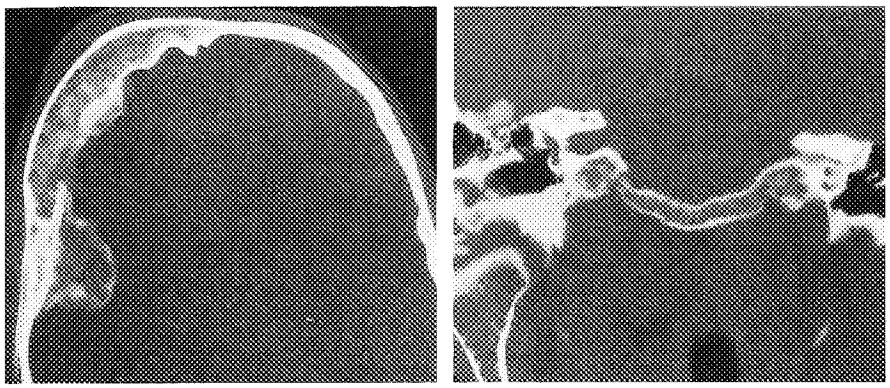
Fig. 5.—44-year-old woman who initially had polyostotic fibrous dysplasia at age 12.

***A*, Axial CT image shows complete stenosis of right external canal due to bony overgrowth.**

***B*, Slightly lower axial section shows involvement of temporomandibular joint with bony overgrowth and secondary degenerative changes.**

***C*, Axial image at level of ventricles shows extension of this process to frontal bone with displacement of anterior frontal and temporal lobes.**

***D*, Coronal image through epitympanic recess shows normal middle ear structures. Note appearance of external canal.**



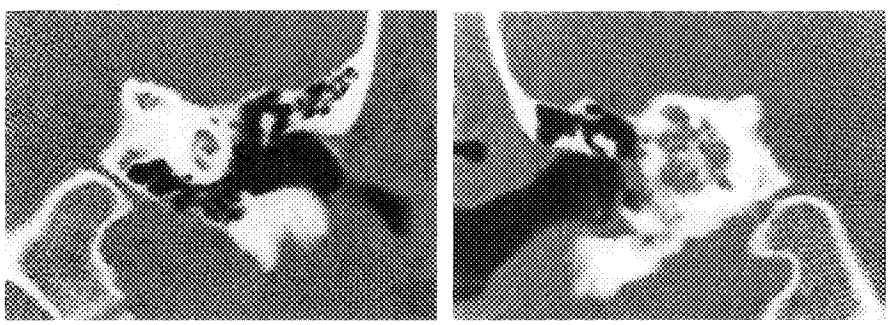


Fig. 6.—12-year-old boy who initially presented at age of 2 years with right external canal mass and 50-dB hearing loss. Mastoidectomy with resection of external canal mass was performed at that time. Images are from 10 years later when patient had severe right-sided sensorineural hearing loss and ipsilateral facial paralysis.

***A*, Coronal CT slice through left cochlea shows normal appearance of left inner ear.**

***B*, Coronal image through right otic capsule shows marked involvement of cochlea.**

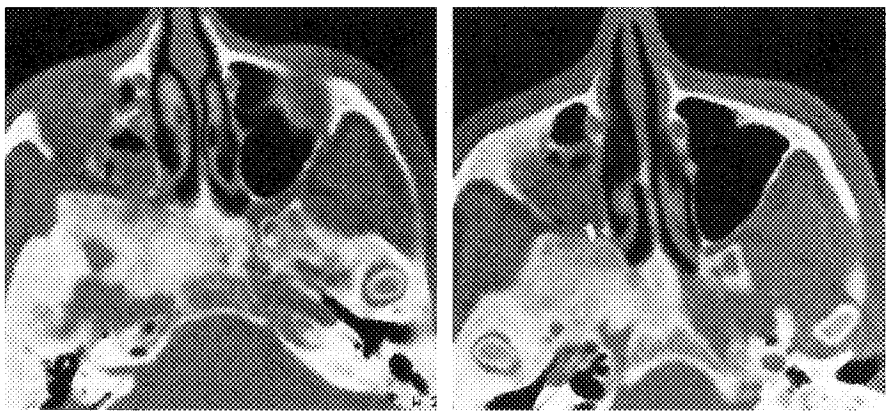


Fig. 7.—33-year-old woman who had headache and right-sided facial pain.

***A*, Axial CT section at level of right internal auditory canal shows involvement of sphenoid bone, right middle cranial fossa, and temporal bone.**

***B*, Slightly lower axial section shows marked thickening of temporal bone surrounding mandibular condyle. Patient's pain was related primarily to temporomandibular joint dysfunction.**

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